General

Guideline Title

Practice guideline summary: sudden unexpected death in epilepsy incidence rates and risk factors: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society.

Bibliographic Source(s)

Harden C, Tomson T, Gloss D, Buchhalter J, Cross JH, Donner E, French JA, Gil-Nagel A, Hesdorffer DC, Smithson WH, Spitz MC, Walczak TS, Sander JW, Ryvlin P. Practice guideline summary: sudden unexpected death in epilepsy incidence rates and risk factors: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Neurology. 2017 Apr 25;88(17):1674-80. [40 references] PubMed

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Recommendations

Major Recommendations

Definitions of the levels of the recommendations (A, B, C, U) and classification of the evidence (Class I-IV) are provided at the end of the "Major Recommendations" field.

Question 1: What Is the Incidence of Sudden Unexpected Death in Epilepsy (SUDEP) in Different Epilepsy Populations?

Incidence Recommendation 1: SUDEP Incidence in Children

Clinicians caring for children with epilepsy should inform the children's parents or guardians that (Level B for the following):

1. There is a rare risk of SUDEP.
2. In 1 year, SUDEP typically affects 1 in 4,500 children with epilepsy; in other words, annually, 4,499 of 4,500 children will not be affected by SUDEP.

Incidence Recommendation 2: SUDEP Incidence in Adults

Clinicians should inform adult persons with epilepsy that (Level B for the following):

1. There is a small risk of SUDEP.
2. In 1 year, SUDEP typically affects 1 in 1,000 adults with epilepsy; in other words, annually, 999 of 1,000 adults will not be affected by

SUDEP.

Question 2: Are There Any Risk Factors for SUDEP?

Recommendation 3

For persons with epilepsy who continue to experience generalized tonic-clonic seizures (GTCS), clinicians should continue to actively manage epilepsy therapies to reduce seizure occurrences and the risk of SUDEP while incorporating patient preferences and weighing the risks and benefits of any new approach (Level B).

Recommendation 4

For persons with frequent GTCS and nocturnal seizures, clinicians may advise selected patients and families, if permitted by their individualized epilepsy and psychosocial circumstances, to use nocturnal supervision or other nocturnal precautions, such as the use of a remote listening device, to reduce SUDEP risk (Level C).

Recommendation 5

Clinicians should inform patients with epilepsy that seizure freedom, particularly freedom from GTCS (which is more likely to occur with medication adherence), is strongly associated with a decreased risk of SUDEP (Level B).

Additional Conclusions (No Recommendations Made)

The evidence is low that the following factors are associated with altering SUDEP risk:

Nocturnal seizures (associated with increased risk)
Any specific antiepileptic drug (AED) (none associated specifically with increased risk)
Lamotrigine use in women (associated with increased risk)
Never having been treated with an AED (associated with increased risk)
Number of AEDs used overall (associated with increased risk)
Heart rate variability (not associated with increased risk)
Extratemporal epilepsy (associated with increased risk)
Intellectual disability (associated with increased risk)
Male sex (associated with increased risk)
Anxiolytic drug use (associated with increased risk)

The evidence is very low or conflicting that the following factors are associated with altering SUDEP risk:

1. Overall seizure frequency when evaluated by using all seizure types
2. Medically refractory epilepsy vs not having well-controlled seizures defined as no seizures in the last year
3. Monotherapy vs polytherapy
4. Carbamazepine, phenytoin, or sodium valproate levels that are above, below, or within the reference range
5. Psychotropic drug use
6. Mental health disorders, lung disorders, or alcohol use
7. Lamotrigine use in people with highly refractory epilepsy
8. Frequent changes in AEDs
9. Therapeutic drug monitoring
10. Undergoing a resective epilepsy surgical procedure (although current research does not rule out the possibility of a beneficial effect or, further, the potential effect of epilepsy surgery on reducing GTCS frequency and epilepsy severity on reducing SUDEP risk)
11. Engel outcome of epilepsy surgery (although current research does not rule out the possibility of a beneficial effect and, further, the potential effect of epilepsy surgery on reducing GTCS frequency and epilepsy severity on reducing SUDEP risk)
12. Vagus nerve stimulator use for more than 2 years (however, current research does not rule out the possibility of a beneficial effect and, further, the potential effect of epilepsy surgery on reducing GTCS frequency and epilepsy severity on reducing the risk of SUDEP)
13. Epilepsy etiology, whether idiopathic or localization-related
14. Structural lesion on magnetic resonance imaging (MRI)
15. Duration of epilepsy
16. Age at epilepsy onset
17. Postictal electroencephalogram (EEG) suppression

Definitions

Rules for Classification of Evidence for Risk of Bias

Screening Scheme

Class I

A statistical, population-based sample of patients studied at a uniform point in time (usually early) during the course of the condition. All patients undergo the intervention of interest. The outcome, if not objective, is determined in an evaluation that is masked to the patients' clinical presentations.

Class II

A statistical, non-referral-clinic-based sample of patients studied at a uniform point in time (usually early) during the course of the condition. Most patients undergo the intervention of interest. The outcome, if not objective, is determined in an evaluation that is masked to the patients' clinical presentations.

Class III

A sample of patients studied during the course of the condition. Some patients undergo the intervention of interest. The outcome, if not objective, is determined in an evaluation by someone other than the treating physician.

Class IV

Studies not meeting Class I, II, or III criteria, including consensus, expert opinion, or a case report.

Prognostic Accuracy Scheme

Class I

A cohort study of a broad spectrum of persons at risk for developing the outcome (e.g., target disease, work status). The outcome is defined by an acceptable reference standard for case definition. The outcome is objective or measured by an observer who is masked to the presence of the risk factor. Study results allow calculation of measures of prognostic accuracy.

Class II

A case-control study of a broad spectrum of persons with the condition compared with a broad spectrum of controls, or a cohort study of a broad spectrum of persons at risk for the outcome (e.g., target disease, work status) where the data were collected retrospectively. The outcome is defined by an acceptable reference standard for case definition. The outcome is objective or measured by an observer who is masked to the presence of the risk factor. Study results allow calculation of measures of prognostic accuracy.

Class III

A case-control study or a cohort study where either the persons with the condition or the controls are of a narrow spectrum where the data were collected retrospectively. The outcome is defined by an acceptable reference standard for case definition. The outcome is objective or measured by an observer who did not determine the presence of the risk factor. Study results allow calculation of measures of a prognostic accuracy.

Class IV

Studies not meeting Class I, II, or III criteria, including consensus, expert opinion, or a case report.

Classification of Recommendations

Assigning a Level of Strength to the Recommendation

When there is sufficient evidence to support an inference for the use of an intervention (i.e., the balance of benefits and harms favors the intervention), the author panel assigns one of three recommendation designations: A, B, or C. Each designation corresponds to a helping verb that denotes the level of strength of the recommendation. Level A is the strongest recommendation level and is denoted by the use of the helping verb *must*. *Must* recommendations are rare, as they are based on high confidence in the evidence and require both a high magnitude of benefit and low risk. Level B corresponds to the helping verb *should*. *Should* recommendations tend to be more common, as the requirements are less stringent but still based on the evidence and benefit–risk profile. Finally, Level C corresponds to the helping verb *may* or *might*. *May* and *might*

recommendations represent the lowest allowable recommendation level the American Academy of Neurology (AAN) considers useful within the scope of clinical practice and can accommodate the highest degree of practice variation.

Level A denotes a practice recommendation that "must" be done. In almost all circumstances, adherence to the recommendation will improve health-related outcomes. A Level B indicates a recommendation that "should" be done. In most circumstances, adherence to the recommendation will likely improve health-related outcomes. A Level C represents a recommendation that "might" be done. In some circumstances, adherence to the recommendation might improve health-related outcomes.

When there is insufficient evidence to support an inference for the use of an intervention (i.e., the balance of benefits and harms is unknown) a Level U or Level R designation is appropriate.

A Level U indicates that the available evidence is insufficient to support or refute the efficacy of an intervention. A Level R is assigned when the balance of benefits and harms is unknown and the intervention is known to be expensive or have important risks. A Level R designates that the intervention should not be used outside of a research setting. Non-evidence-based factors that need to be transparently and systematically considered when formulating recommendations include the following:

- The relative value of the benefit as compared with the risk; this is derived from consideration of:
  - The importance to patients of the health related-outcomes (both benefits and harms)
  - The size of the intervention's effect
  - The risk of harm of the intervention (i.e., tolerability and safety)
- The feasibility of complying with the intervention (e.g., the intervention's availability)
- The cost of the intervention
- The expected variation in patient preferences relative to the risks, burdens, and benefits of the intervention

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

- Sudden unexpected death in epilepsy (SUDEP)
- Generalized tonic-clonic seizures (GTCS)
- Epilepsy

Guideline Category

Counseling

Management

Risk Assessment

Clinical Specialty

Internal Medicine

Neurology

Pediatrics

Intended Users

Advanced Practice Nurses

Health Care Providers

Patients

Physician Assistants

Physicians

Guideline Objective(s)

- To examine evidence for the sudden unexpected death in epilepsy (SUDEP) incidence rate in epilepsy populations and for prognostic factors for SUDEP occurrence
- To inform an honest and balanced discussion when clinicians counsel people about SUDEP, and provide insight into areas where more clinical research is needed

Target Population

Adults and children with epilepsy

Interventions and Practices Considered

1. Informing adults and parents/guardians of children with epilepsy of risk and incidence of sudden unexpected death in epilepsy (SUDEP)
2. Active management of epilepsy therapies in persons who continue to experience generalized tonic-clonic seizures (GTCS), including incorporating patient preferences and weighing risks and benefits of any new approach
3. Advising patients and families to use nocturnal supervision or other nocturnal precautions (e.g., remote listening device) for patients with frequent GTCS and nocturnal seizures
4. Informing patients with epilepsy that seizure freedom is strongly associated with a decreased risk of SUDEP

Major Outcomes Considered

- Incidence of sudden unexpected death in epilepsy (SUDEP)
- Risk factors for SUDEP

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

In 2010, the American Academy of Neurology (AAN) Guideline Development, Dissemination, and Implementation Subcommittee (GDDI) and the Guidelines Committee of the American Epilepsy Society convened a panel of experts to develop this practice guideline. In November 2010, an independent librarian performed a systematic literature search of MEDLINE and EMBASE in all languages from earliest available article to November 2010. The guideline panel performed an identical search in April 2015 to include articles published since November 2010. The following keywords were used in both the 2010 and the 2015 searches: SUDEP OR (sudden AND (unexplained OR unexpected) AND death). These were combined with the traditional medical subheadings (MeSH) for epilepsy ("Epilepsy/abnormalities" OR "Epilepsy/classification" OR "Epilepsy/complications" OR "Epilepsy/drug effects" OR "Epilepsy/drug therapy" OR "Epilepsy/epidemiology" OR "Epilepsy/ethnology" OR "Epilepsy/etiology" OR "Epilepsy/genetics" OR "Epilepsy/mortality" OR "Epilepsy/physiopathology" OR "Epilepsy/prevention and control" OR

"Epilepsy/therapy") with limits of "Humans," plus "All Child: 0–18 years" or "All Adult: 19+ years." Literature types were limited to "Clinical Trial, Randomized Controlled Trial, Comparative Study, Controlled Clinical Trial, Evaluation Studies, Journal Article, Multicenter Study, Research Support, N I H, Extramural, Research Support, N I H, Intramural, Research Support, Non U S Gov't, Research Support, U S Gov't, Non P H S, Research Support, U S Gov't, P H S, Validation Studies." Finally, the guideline panel specifically searched causes implicated in SUDEP (e.g., cardiac arrhythmias and preictal autonomic dysfunction) where the hypotheses were tested. See appendix e-4 in the online Data Supplement for the complete search strategy.

Two panel members working independently of each other reviewed each of the resulting 1,068 abstracts to establish whether any of the corresponding articles met the inclusion criteria (data relevant to questions, cohort, case control, case series, n >10). A total of 744 abstracts were excluded at this point because the corresponding articles did not include data that addressed either question, such as not addressing actual SUDEP occurrences but evaluating possible SUDEP risk factors. A total of 324 abstracts met criteria for full-text review, and their corresponding articles were reviewed. Of those, 70 articles met criteria for classification and each was classified by at least 2 GDDI committee panel members reviewing independently of each other. Reviewed articles were entered into a database application through an online questionnaire. Thirty-five articles had data for inclusion. The remaining articles were excluded because they did not have data that addressed the question or otherwise did not meet inclusion criteria, did not employ an adequate sudden unexpected death in epilepsy (SUDEP) definition, or did not use an appropriate epilepsy comparison group in the prognostic studies.

Included articles were required to state that the sudden unexpected death in epilepsy (SUDEP) definition provided by Nashef 1997, Annegers 1997, and Leestma et al. 1997 was used or to describe criteria in accordance with these definitions. These definitions share the following criteria: (1) Patients had epilepsy by reasonable criteria. (2) Deaths by drowning, trauma, or status epilepticus were excluded. (3) Death could have occurred after a witnessed seizure. (4) Other competing causes of death were excluded.

Number of Source Documents

Thirty-five articles had data for inclusion.

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Rules for Classification of Evidence for Risk of Bias

Screening Scheme

Class I

A statistical, population-based sample of patients studied at a uniform point in time (usually early) during the course of the condition. All patients undergo the intervention of interest. The outcome, if not objective, is determined in an evaluation that is masked to the patients' clinical presentations.

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Prognostic Accuracy Scheme

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A case-control study or a cohort study where either the persons with the condition or the controls are of a narrow spectrum where the data were collected retrospectively. The outcome is defined by an acceptable reference standard for case definition. The outcome is objective or measured by an observer who did not determine the presence of the risk factor. Study results allow calculation of measures of a prognostic accuracy.

Class IV

Studies not meeting Class I, II, or III criteria, including consensus, expert opinion, or a case report.

Methods Used to Analyze the Evidence

Meta-Analysis

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

The guideline panel used 2 of the American Academy of Neurology's (AAN's) evidence-based schemes to rate articles: the screening criteria for the incidence question and the prognostic criteria for the risk factor question (see the "Rating Scheme for the Strength of the Evidence" field). Strong evidence for incidence criteria was supported by a population-representative cohort of people with epilepsy evaluated by either prospective or retrospective methods (retrospective accepted because of the objective nature of the outcome assessed), more than 80% completeness of evaluation of deaths within that cohort, and more than 80% evaluation of those deaths for sudden unexpected death in epilepsy (SUDEP) as a cause. In these articles, the total number of patient-years of follow-up must have been provided or readily derived in order to calculate the denominator for SUDEP rates. Strong evidence for SUDEP risk factors came from studies that prospectively followed a cohort of patients with epilepsy and compared the frequency with which factors were present in cases of SUDEP vs in living persons with epilepsy. Retrospective case-control studies provided the majority of the data regarding SUDEP risk factors (Class II evidence). Because of the variability in the completeness of data in SUDEP research, SUDEP cases are categorized as definite, probable, and possible. Definite SUDEP cases meet the criteria stated previously and have a postmortem report. Probable SUDEP cases meet all the criteria stated previously but lack postmortem data. Possible SUDEP cases are those for which SUDEP cannot be ruled out but there is insufficient evidence regarding the circumstances of death, potential competing causes of death (such as presence of cardiac risk factors), and lack of an available postmortem report. Only definite and probable cases were included in this analysis.

The available literature consisted of multiple Class I articles for incidence, and therefore articles rated Class II or lower for incidence were excluded. Several Class I and many more Class II articles were available for prognostic questions. For the 2 included articles published before the accepted SUDEP definition from 1997 was established, the definition was still applied.

Strength of the evidence to determine final conclusions was derived according to a modified Grading Recommendations Assessment, Development and Evaluation (GRADE) process. This modified GRADE process requires that the guideline panel reach agreement on whether to downgrade the evidence because of a lack of precision, consistency, generalizability, or biological plausibility, or to upgrade the evidence because of a clear dose response or a large magnitude of effect. Random-effects meta-analyses were performed for incidence studies of similar populations in order to derive summary measures when confidence intervals (CIs) for these studies were dissimilar.

Methods Used to Formulate the Recommendations

Expert Consensus (Delphi)

Description of Methods Used to Formulate the Recommendations

The recommendations were first anchored in the strength of the conclusions and then further modulated by application of a modified Delphi process. Through this process, the recommendation levels could be dissociated to some degree from the evidence strength. For example, recommendations could be downgraded on the basis of drawbacks such as cost, patient preference, or availability of the intervention, or upgraded because of a low degree of risk in relation to benefit. Many conclusions were of sufficient strength to support counseling recommendations. Although the therapeutic classification scheme was not used because the literature search did not find any treatment trials, therapeutic recommendations were derived from risk factors that could be modified by medical intervention. The level of the therapeutic recommendation was anchored in the strength of the conclusion regarding the associated prognostic risk factor and was subject to downgrading or upgrading as just described.

Rating Scheme for the Strength of the Recommendations

Classification of Recommendations

Assigning a Level of Strength to the Recommendation

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Level A denotes a practice recommendation that "must" be done. In almost all circumstances, adherence to the recommendation will improve health-related outcomes. A Level B indicates a recommendation that "should" be done. In most circumstances, adherence to the recommendation will likely improve health-related outcomes. A Level C represents a recommendation that "might" be done. In some circumstances, adherence to the recommendation might improve health-related outcomes.

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A Level U indicates that the available evidence is insufficient to support or refute the efficacy of an intervention. A Level R is assigned when the balance of benefits and harms is unknown and the intervention is known to be expensive or have important risks. A Level R designates that the intervention should not be used outside of a research setting. Non-evidence-based factors that need to be transparently and systematically considered when formulating recommendations include the following:

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  - The importance to patients of the health related-outcomes (both benefits and harms)
  - The size of the intervention's effect
  - The risk of harm of the intervention (i.e., tolerability and safety)
- The feasibility of complying with the intervention (e.g., the intervention's availability)
- The cost of the intervention
- The expected variation in patient preferences relative to the risks, burdens, and benefits of the intervention

Cost Analysis

A formal cost analysis was not performed, and published cost analyses were not reviewed.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

Drafts of the practice guidelines have been reviewed by at least 3 American Academy of Neurology (AAN) committees, at least 1 American Epilepsy Society (AES) committee, a network of neurologists, *Neurology*® peer reviewers, and representatives from related fields.

This guideline was approved by the Guideline Development, Dissemination, and Implementation Subcommittee on November 7, 2015; by the AAN Practice Committee on January 17, 2016; by the AES Guidelines Committee on November 11, 2016; by the AES Council on Clinical Activities on November 11, 2016; by the AES Executive Committee on November 14, 2016; by the AES Board of Directors on November 30, 2016; and by the AAN Institute Board of Directors on January 11, 2017. This practice guideline was endorsed by the International Child Neurology Association on August 27, 2016.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

- It seems reasonable to infer that improved control of an individual's generalized tonic-clonic seizures (GTCS) will result in a reduced risk of sudden unexpected death in epilepsy (SUDEP). Thus, a reduction in SUDEP risk is an additional benefit to the many benefits resulting from improved seizure control.
- People with epilepsy and their families prefer to be informed of the individual's risk for a catastrophic event such as SUDEP, even when the probability of the event is low. After being informed of an adverse event, people commonly overestimate the risk of that adverse event happening to them. Such overestimation unduly increases anxiety related to an adverse event. Overestimation can be lessened by presenting the risk as the probability of both having and not having the event, and by using numbers in addition to words and frequencies rather than percentages to convey the risk.
- The presence in the bedroom of another individual at least 10 years of age and of normal intelligence is associated with a decreased SUDEP risk. If it were in accordance with patient and family circumstances and values, nocturnal supervision could reduce SUDEP risk.
- Patients are especially interested in factors that might reduce their risk even when a causal link between the factor and a reduction in risk has not been established. Knowledge of these risk factors might suggest behaviors that could modify the risk factors (e.g., improved therapy adherence), increase the person's sense of control, and reduce the anxiety that comes from awareness of the risk.

Potential Harms

- As with all benefits associated with improved seizure control, the potential benefit of sudden unexpected death in epilepsy (SUDEP) risk reduction needs to be balanced with the risks and burdens associated with antiseizure therapies.
- If it were in accordance with patient and family circumstances and values, nocturnal supervision could SUDEP risk; however, providing nighttime observation might be overly burdensome and intrusive.

Qualifying Statements

Qualifying Statements

Disclaimer

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Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Patient Resources

Quick Reference Guides/Physician Guides

Resources

Slide Presentation

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

Harden C, Tomson T, Gloss D, Buchhalter J, Cross JH, Donner E, French JA, Gil-Nagel A, Hesdorffer DC, Smithson WH, Spitz MC, Walczak TS, Sander JW, Ryvlin P. Practice guideline summary: sudden unexpected death in epilepsy incidence rates and risk factors: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Neurology. 2017 Apr 25;88(17):1674-80. [40 references] PubMed

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2017 Apr 25

Guideline Developer(s)

American Academy of Neurology - Medical Specialty Society

American Epilepsy Society - Disease Specific Society

Source(s) of Funding

This guideline was developed with financial support from the American Academy of Neurology (AAN). Authors who serve as AAN subcommittee members or methodologists were reimbursed by the AAN for expenses related to travel to subcommittee meetings where drafts of manuscripts were reviewed.

Guideline Committee

Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology

American Epilepsy Society Guidelines and Assessment Committee

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Financial Disclosures/Conflicts of Interest

Conflicts of Interest

The American Academy of Neurology (AAN) and the American Epilepsy Society (AES) are committed to producing independent, critical, and truthful practice guidelines. Significant efforts are made to minimize the potential for conflicts of interest to influence the recommendations of this practice guideline. To the extent possible, the AAN and the AES keep separate those who have a financial stake in the success or failure of the products appraised in the practice guidelines and the developers of the practice guidelines. Conflict of interest forms were obtained from all authors and reviewed by an oversight committee prior to project initiation. AAN and AES limit the participation of authors with substantial conflicts of interest. The AAN and the AES forbid commercial participation in, or funding of, practice guidelines projects.

Disclosures

C. Harden has received royalties from Wiley and UpToDate and has served as a contributing editor for *Epilepsy Currents*. T. Tomson has served as the associate editor of *Epilepsia*; is a member of the editorial boards of *Epilepsy Research*, *Epileptic Disorders*, and the *European Journal of Clinical Pharmacology*; has received honoraria from Sun Pharmaceuticals, UCB, Eisai, and Bial; has served as a member of an expert panel for sudden unexpected death in epilepsy (SUDEP) adjudication in clinical trials of lamotrigine sponsored by GlaxoSmithKline; and has received research support from UCB, GlaxoSmithKline, Bial, Eisai, Novartis, Stockholm County Council, and Citizens United in Research for Epilepsy (CURE). D. Gloss serves as an evidence-based medicine consultant for the American Academy of Neurology (AAN) and has served as an associate editor (risk of bias classification) for *Neurology®*. J. Buchhalter has received funding for travel from the AAN; serves on an editorial advisory board for *Pediatric Neurology* and *Epilepsy Currents*; has served as a consultant to UCB, Upsher-Smith Laboratories, and Eisai; and has performed clinical procedures/imaging studies related to the content of this practice guideline, including EEG and video EEG (25%) and epilepsy surgery evaluation. J. Cross has served as a member of the editorial boards of *Developmental Medicine*, *Child Neurology*, and the *European Journal of Child Neurology*; has a patent for C10 in the treatment of epilepsy; has received royalties for a chapter on childhood epilepsy in *Brain Diseases of the Nervous System* and as editor of *Paediatric Epilepsy*; has received research support from the UK National Institute for Health and Research (NIHR), the European Framework FP7, the Charles Wolfson Foundation, Action Medical Research, and Sparks; and has sat on advisory boards for Vitaflo, Sanofi, Eisai, Viropharma, and Zogenix, for which remuneration is paid to her department. E. Donner has received research support from the Canadian Institutes of Health Research, Dravet Canada, and SUDEP Aware. J. French has served as a consultant for Acorda, Biotie, Eisai Medical Research, GlaxoSmithKline, Impax, Johnson & Johnson, Lewis County General Hospital, Marinus, Novartis, Pfizer, Sunovion, SK Life Science, Supernus Pharmaceuticals, UCB, Upsher-Smith, and Vertex; has received grants from Eisai Medical Research, the US Epilepsy Research Foundation, the Epilepsy Study Consortium, the Epilepsy Therapy Project of the Epilepsy Foundation, Lundbeck, Pfizer, and UCB; and is president of the Epilepsy Study Consortium. All consulting is done on behalf of the Consortium, and fees are paid to the Consortium. New York University receives salary support from the Consortium. A. Gil-Nagel has received personal compensation from Bial, Eisai, GSD Pharma Consulting, UCB Pharma, and Pfizer; has received funding for travel from Bial, Eisai, and GlaxoSmithKline; has served as an editor for *Seizure*, *Neurologia*, and *Revista de Neurologia*; has served on speakers bureaus for Bial, Eisai, GlaxoSmithKline, and UCB Pharma; and asserts that the information he provides his patients in his epilepsy clinic may be influenced by the results of this practice guideline. D. Hesdorffer is a member of the SUDEP Institute and of the Executive Committee of the North American SUDEP Registry; has served on scientific advisory boards for Upsher-Smith and Acorda; has served as a consultant for Cyberonics; has received funding for travel from the International League Against Epilepsy; has served as an associate editor of *Epilepsia*; has served on the editorial board for *Epilepsy and Behavior*; has served as a contributing editor for *Epilepsy Currents*; and has received funding from the NIH, the Centers for Disease Control and Prevention, the Epilepsy Consortium, the Patient Centered Outcome Research Institute, Finding a Cure for Epilepsy, The Epilepsy Study Consortium, and the Icahn School of Medicine at Mount Sinai (for consulting work on an injury prevention grant). W. Smithson has served on a scientific advisory board for the Sanofi UK consensus guidelines on women with epilepsy, has received travel funding for the Partners Against Mortality in Epilepsy conference on SUDEP (Washington 2016), has received publishing royalties from Blackwell Publishing for the *ABC of Epilepsy*, has received financial support in the form of funding for a general practice research infrastructure from the NIHR (UK), and has given expert witness testimony for the Fatal Accident Inquiry Dundee 2012 (2 cases of SUDEP). M. Spitz has received personal compensation and honoraria for serving on an advisory board for UCB, has received travel funding from Cyberonics (to see the site/factory), has received financial support for a U.S. Department of Defense Study on closed head injury, and has given expert testimony, prepared an affidavit for, and acted as a witness or consultant regarding a legal proceeding. T. Walczak serves on a scientific advisory panel tracking incidence of SUDEP in follow-up of patients treated with the NeuroPace RNS System. Compensation goes directly to his academic department and does not increase his salary. J. Sander is based at University College London/University College London Hospitals, which receives funding from the UK Department of Health's NIHR Biomedical Research Centres; has served on advisory boards for UCB and Eisai; has received speaker honoraria from GlaxoSmithKline, Eisai, UCB, Lundbeck, and Teva; serves on the editorial board of the *Lancet Neurology*; and receives research support from the Dr. Marvin Weil Epilepsy Research Fund, the Epilepsy Society (UK), the Netherlands Epilepsy Fund, Eisai, GlaxoSmithKline, WHO, and EU FP7. His current position is endowed by the Epilepsy Society (UK). P. Ryvlin has served as a chair of the Scientific Advisory Committee for the annual meeting of the French League Against Epilepsy; has received travel funding and honoraria from GlaxoSmithKline, Eisai, Janssen Cilag Pty. Ltd., Cyberonics, Medtronic, and UCB Pharma (in order to participate on industry-funded advisory boards or symposia); has served as

a journal editor for *Epilepsia*, *Epilepsy Research*, *Epileptic Disorders*, and *Epilepsy Research and Treatment*; has served on speakers bureaus for Eisai, GlaxoSmithKline, and UCB Pharma for a symposium at the European and International Epilepsy Congress (in order to participate on advisory boards or symposia); and has received financial support in the form of a European FP7 grant (EURIPIDES) and grant/research program funding from national (French) entities, including 2 PHRC (Programme Hospitalier de Recherche Clinique), 1 INSERM-DHOS (Institut National de la Santé et de la Recherche Médicale-Direction de l'Hospitalisation et de l'Organisation des Soins) Translationnelle, and 1 Contrat d'Interface INSERM. Go to Neurology.org for full disclosures.

Guideline Endorser(s)

International Child Neurology Association - Medical Specialty Society

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Guideline Availability

A list of American Academy of Neurology (AAN) guidelines, along with a link to this guideline, is available from the AAN Web site.

Availability of Companion Documents

The following are available:

- Practice guideline: sudden unexpected death in epilepsy incidence rates and risk factors. Summary of practice guideline for clinicians. Minneapolis (MN): American Academy of Neurology; 2017 Apr. 3 p. Available from the American Academy of Neurology (AAN) Web site.
- Harden C, Tomson T, Gloss D, Buchhalter J, Cross JH, Donner E, French JA, Gil-Nagel A, Hesdorffer DC, Smithson WH, Spitz MC, Walczak TS, Sander JW, Ryvlin P. Practice guideline: sudden unexpected death in epilepsy incidence rates and risk factors: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Data supplement. Minneapolis (MN): American Academy of Neurology; 2017 Apr. Available from the Neurology Web site.
- Practice guideline: sudden unexpected death in epilepsy incidence rates and risk factors: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Presentation slides. Minneapolis (MN): American Academy of Neurology; 2017 Apr. 44 p. Available from the AAN Web site.
- Practice guideline summary: sudden unexpected death in epilepsy incidence rates and risk factors: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Video of press conference held in Boston, MA. Available from the AAN Web site.
- Practice guideline summary: sudden unexpected death in epilepsy incidence rates and risk factors: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Podcast. Available from the Neurology Web site.
- American Academy of Neurology (AAN). Clinical Practice Guideline Process Manual, 2011 Ed. Minneapolis (MN): American Academy of Neurology. Available from the AAN Web site.

Patient Resources

The following are available:

- Sudden unexpected death in epilepsy incidence rates and risk factors. AAN summary of practice guideline for patients and their families.

Minneapolis (MN): American Academy of Neurology; 2017. 1 p. Available from the American Academy of Neurology (AAN) Web site .

- Video for patients. SUDEP. Available from the AAN Web site .
- Public service announcement. Available from the AAN Web site .

Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

NGC Status

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